Biphasic Amyloidosis Responsive to Acitretin

Asitetine Cevap Veren Bifazik Amiloidoz

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Özet

Primer kutanöz amiloidoz başka bir deri veya sistemik hastalığı olmaksızın, deride amiloid birikimi ile karakterize bir grup hastalıktır. Maküler ve liken amiloidozun bir arada görülmesi "bifazik amiloidoz" olarak adlandırılır. Yirmi bir yaşında erkek hasta polikliniğimize göğüste hiperpigmentasyon, özellikle bacaklarında yoğunlaşan kaşıntılı hiperkeratotik papüller şikayeti ile başvurdu. Hastaya bifazik amiloidoz ve bronşial astım tanısı kondu. Hastaya 6 ay boyunca oral acitretin tedavisi verildi. Tedavi sonrasında kaşıntı ve papüllerinde remisyon izlendi. Bildiğimiz kadarıyla; bu olgu asitretin ile başarılı bir şekilde tedavi edilen ve bronşial astımın eşlik ettiği ilk bifazik amiloidoz vakasıdır.

Anahtar Kelimeler

Bifazik Amiloidoz; Bronşial Astım; Asitretin

Abstract

Primary cutaneous amyloidosis is a group of diseases characterized by deposition of amyloid in skin without association with other cutaneous or systemic disorders. Coexistence of macular and lichen amyloidosis are termed as "biphasic amyloidosis". A 21-year-old male admitted to our clinic with a hyperpigmentation on chest and intensive pruritic hyperkeratotic papules, mainly located on legs. He diagnosed with biphasic amyloidosis and bronchial asthma. Oral acitretin treatment was given during six months. Remission of papules was noted after the treatment. Best of our knowledge; this was the first biphasic cutaneous amyloidosis case report that was associated with bronchial asthma and successfully treated by acitretin.

Keywords

Biphasic Amyloidosis; Bronchial Asthma; Acitretin

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Introduction

Primary cutaneous amyloidosis is a group of diseases characterized by deposition of amyloid in skin without association with other cutaneous or systemic disorders [1]. Macular amyloidosis and lichen amyloidosis are forms of primary cutaneous amyloidosis. The association of the both types is termed biphasic amyloidosis [2]. In this case report, we present a biphasic amyloidosis associated with bronchial asthma and treated with oral acitretin.

Case Report

A 21-year-old man admitted to our clinic with a hyperpigmentation on chest and intensive pruritic hyperkeratotic papules, mainly located on legs. Cutaneous hyperpigmentation and numerous papules appeared thirteen years ego. Pruritus started when the patient was seventeen years old. Patient did not have family history of skin disorders. It did not recover although it had been treated with topical and/or systemic corticosteroids, systemic antihistamines, and topical emollients by various clinics. Dermatologic examination was revealed extensive grey-brown hyperpigmentation on his back, abdomen, and lumbosacral regions (Figure-1). Numerous hyperkeratotic papules located mainly on

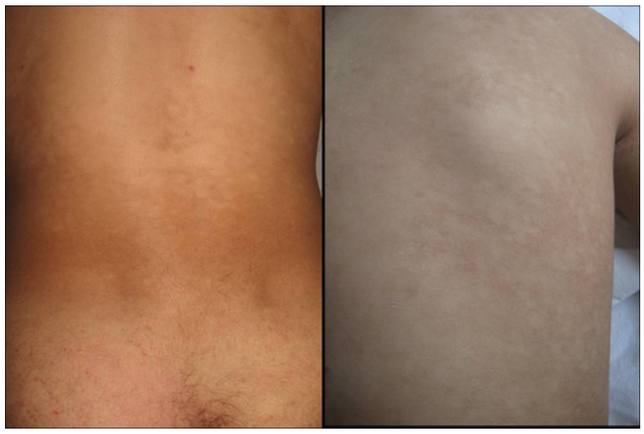

Figure 1. Extensive grey-brown hyperpigmentation on his back, abdomen, lumbosacral regions

the extensor aspects of extremities. The lower extremities were most severely involved by these firm papules (Figure-2). Patient had shortness of breath and a nodule on right pretibial region. The patient underwent chest disease and plastic surgery consultation. Pulmonary function test was performed. The patient diagnosed with bronchial asthma by pulmonologist. The nodule, that was over the right tibia, excised by plastic surgeon and diagnosed with prurigo nodularis. Blood cell count and differential, sedimentation rate, serum calcium, phosphorus, immunoglobulin E (IgE) level, protein electrophoresis, Hbs Ag, Anti-Hbs, Anti-HCV, HIV, hepatic, thyroid, and renal function tests, urinalysis were evaluated and pathology were not found. Thyroid, parathyroid ultrasonography, chest and abdominal X-ray were normal. Examination of the scalp, mucous membranes, palms and soles were normal. Any other abnormality was not found in systemic examination. Biopsy samples were taken from chest and right upper thigh. Histopathological examination of this case lesions revealed orthokeratotic hyperkeratosis, irregular acanthosis, superficial perivascular mononuclear cell infiltration and globular homogeneous eosinophilic deposits stained with crystal violet in the expanded papillary dermis (Figure-3). Microscopic examination was assessed as "macular and lichenoid amyloidosis". Oral acitretin treatment was given during six months. Remission of pruritus and papules were noted after the treatment (Figure-2).

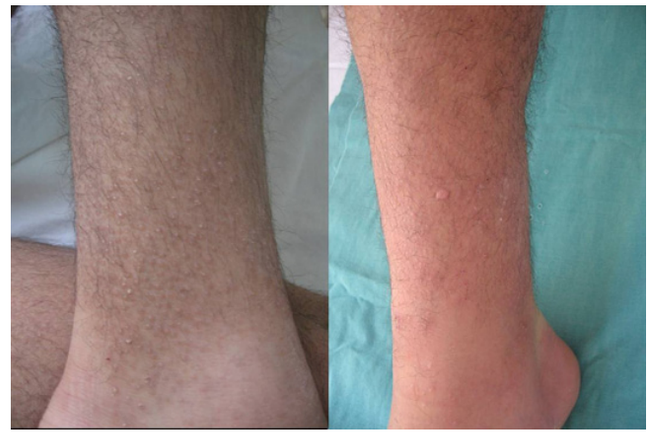

Figure 2. Numerous hyperkeratotic papules located mainly on the extensor aspects of extremities. The papules on the lower extremities became flattened

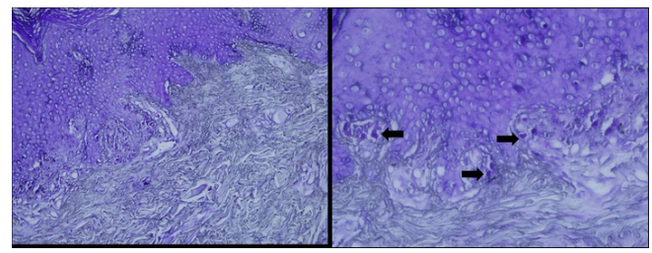

Figure 3. Globular homogeneous eosinophilic deposits that was pointed out with black arrow stained with crystal violet in the expanded papillary dermis (Cristal Violet x200 (left view), x400 (right view)).

Discussion

The amyloidosis is a large group of protein-misfolding diseases. Primary cutaneous amyloidosis (PCA) is deposition of amyloid in previously normal skin, without association with other cutaneous or systemic disorders [1-2]. The association of the macular amyloidosis (MA) and lichen amyloidosis (LA) is termed biphasic amyloidosis (BA) [2]. Numerous brownish lichenoid papules are seen in LA especially over the extremities. Pigmented macule may be appeared as reticulate or rippled in MA [3]. In this case, extensive reticulated grey-brown hyperpigmentation on his back, abdomen, lumbo-sacral regions, and numerous hyperkeratotic firm papules that located mainly on the extensor aspects of extremities were noted and diagnosed BA.

The pathogenesis of PCA is poorly understood. However, it is generally accepted that, in MA and LA, amyloid occurs mainly as a result of keratinocyte degeneration, and there is no relation with systemic amyloidosis [3]. One proposed mechanism for keratinocyte degeneration is repeated mechanical trauma induced by chronic friction [4].

Histological examinations of both MA and LA are similar with the deposition of amorphous eosinophilic material in dermal papillae that stains with crystal violet and Congo red [4].

Some cases of LA associated with lichen planus, autoimmune thyroiditis, ankylosing spondylitis, HIV and HCV infection, Sipple syndrome and mycosis fungoides have been reported [5]. Cutaneous amyloidosis is often associated with pruritic disorders such as atopic dermatitis and cholestasis. This leaded some authors to suggest that deposition of amyloid is result of

chronic scratching, caused by long-term pruritus [3]. Presented patient had shortness of breath. Pulmonary function test was performed. The patient diagnosed with bronchial asthma by pulmonologist.

Although various treatment modalities including topical and intralesional steroid injections topical dimethyl sulfoxide (DMSO), ultraviolet B (UVB), oral psoralen plus ultraviolet A (PUVA), retinoid and dermabrasion have been described, the results are generally unsatisfactory [1-3,6]. As with other type of treatments, retinoid therapy has also had controversial results and lack of response [1]. In this case, patient had been treated with topical corticosteroids, oral antihistamines, oral corticosteroids and PUVA (three times a week for 4 months) during 13 years however treatment results were unsatisfactory. The patient underwent oral acitretin treatment at dose of 35 mg/day (0.5 mg/kg) for 6 months. The patient was followed-up every 4 week and repeated routine laboratory examination. The papules on the lower extremities became flattened (Figure-2). The treatment was well tolerated. Routine laboratory examination was normal. Mild cheilitis, cutaneous dryness were noted during treatment. No recurrence has been seen for 10 months in the follow up period.

Conclusion

As far as we know, this is the first report describing biphasic cutaneous amyloidosis associated with bronchial asthma and was successfully treated with acitretin.

Competing interests

The authors declare that they have no competing interests.

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